A PAIN IN MY BULLAE!

An Approach to Managing Bullous Pemphigoid

By: Dr. Aleena Fiorotto

OBJECTIVES

1. Describe the pathophysiology of Bullous Pemphigoid
2. Outline possible triggers of Bullous Pemphigoid
3. Describe management of Bullous Pemphigoid
4. Review possible complications of Bullous Pemphigoid and it's treatment

CONFLICTS OF INTEREST

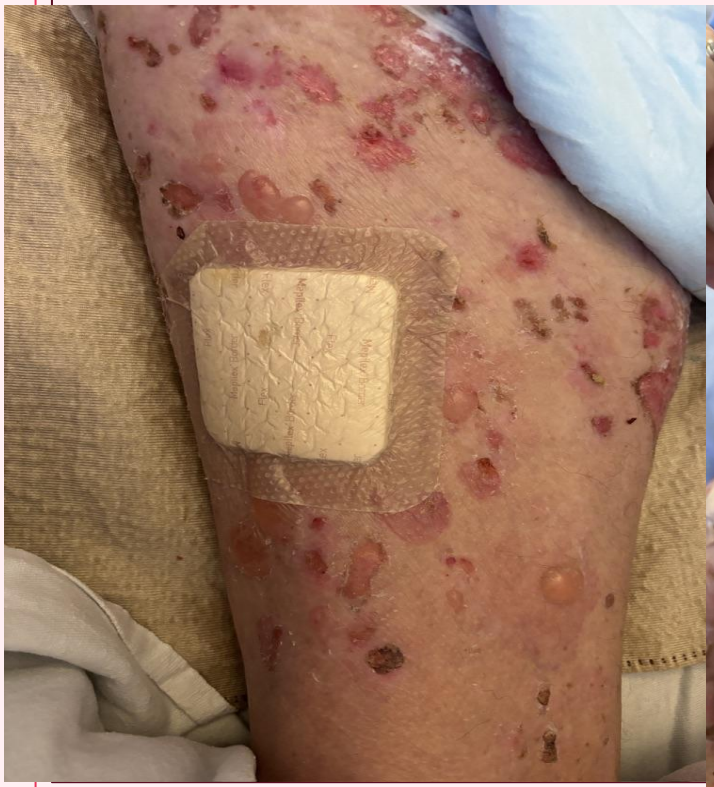
- None
- Dr. Lyne Giroux is my hero, and no, I am not being paid to say this
- I received permission from my patient to use the photos you are about to see.

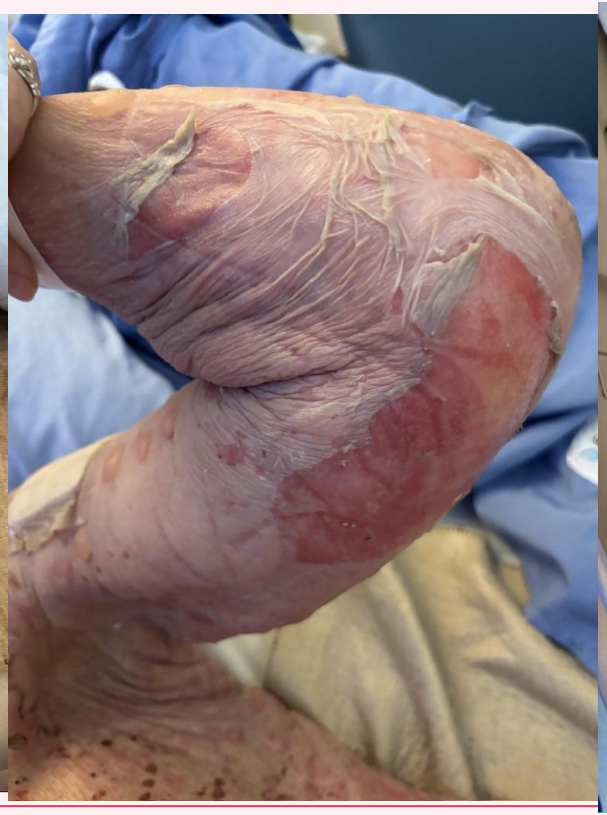
A QUICK CASE:

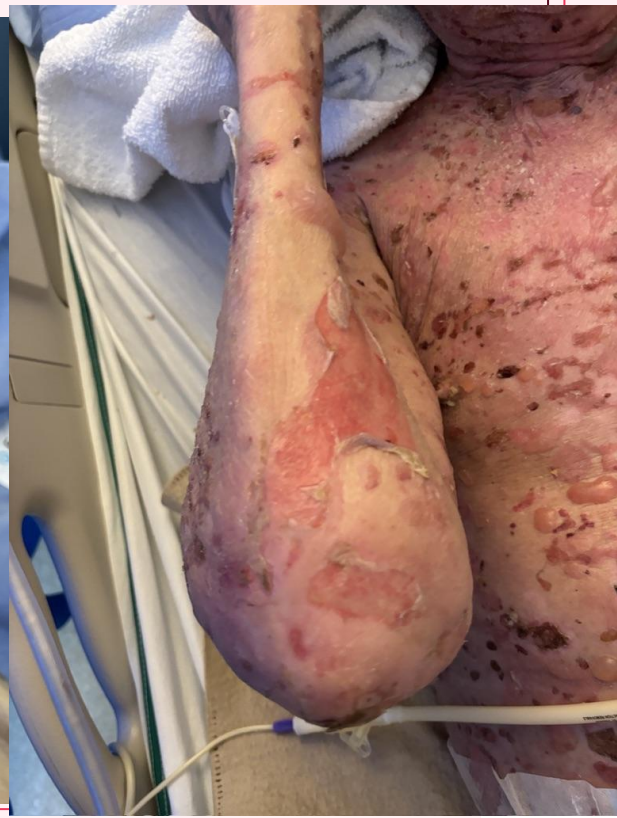
ID: 68 year old male admitted to hospital with aspiration pneumonia + sepsis secondary to LARGE Zenker's diverticulum

- Needed brief ICU Admission and intubation
- Had G-tube placed to await Zenker's repair as was not safe to swallow
- While in hospital: Developed a diffuse bullous rash (apparently had previously the year before, but all lesions had resolved with no treatment)
- Concerned for Bullous Pemphigoid, pt was placed on oral prednisone 80mg daily with plans for taper.

A QUICK CASE







A QUICK CASE:

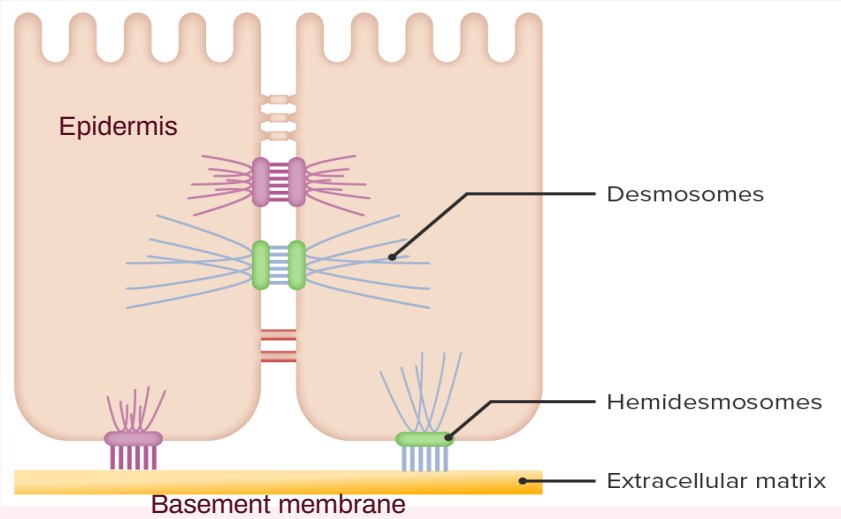
- Despite high-dose oral steroids, no improvement in patient's rash
- Continued to spread over his body, resulting in SIGNIFICANT skin sloughing, hypoalbuminemia, hyponatremia
- Subsequent sepsis (again!) from open wounds/cellulitis
- I needed HELP. Called Dr. Kersti Kents out of desperation and she put me in touch with Dr. Lyne Giroux, a Dermatology-Goddess, based out of Sudbury.
- We will go through her advice later on…

BULLOUS PEMPHIGOID

What is it???

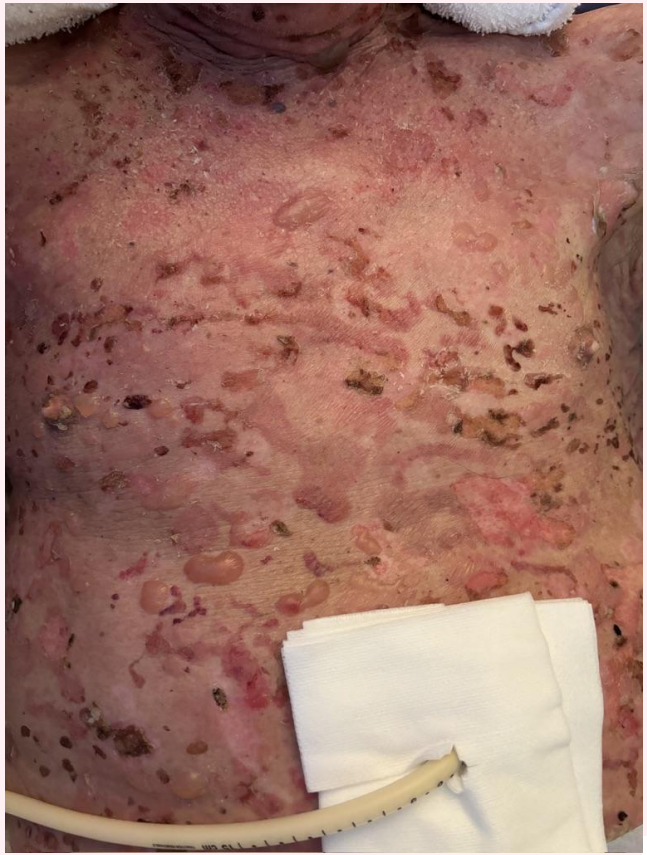
Definition: An autoimmune blistering skin disease characterized by subepidermal bullae

- IgG mediated immune response against hemidesmosomal proteins and basement membrane zone of the skin
- Usually in adults >60 years old
- Most common autoimmune blistering disorder



CLINICAL FEATURES

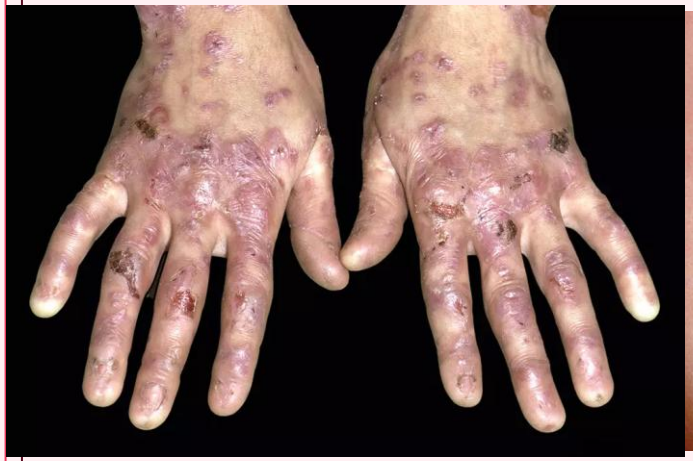
- **Prodromal phase:** pruritus, urticaria-like lesions, eczematous patches
- **Bullous phase:** tense bullae on normal or erythematous skin
- Typically involves trunk, flexor surfaces, abdomen, groin
- Bullae will then rupture→increases risk of secondary infection and scarring



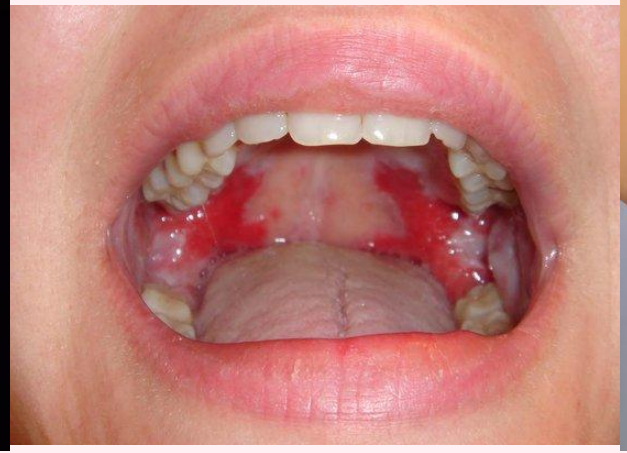
DIAGNOSIS

- Biopsy with direct immunofluorescence showing IgG/C3 along the dermal/epidermal junction
- Special medium required: Michel's medium *** At Bracebridge site, need to ask for it from lab!!!***

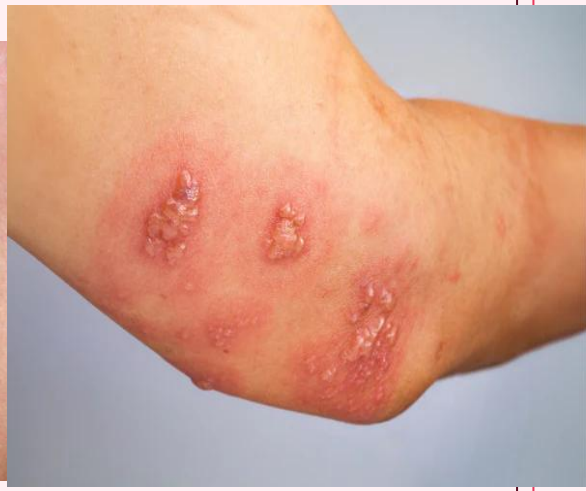
DIFFERENTIAL DIAGNOSIS



Epidermolysis bullosa acquisita
- Deeper layer of skin
- Scarring
- Hand/foot involvement more likely



Pemphigus vulgaris
- Autoantibodies against desmosomes
- flaccid blisters
- mucosal involvement



Dermatitis herpetiformis
- Younger adults
- Itchy clusters of papules/vesicles
- Associated with celiac

OFFENDING MEDICATIONS

- **Gliptins (OZEMPIC!!), hydrochlorothiazide, furosemide, spironolactone, amlodipine, losartan, captopril/lisinopril, clonidine, omeprazole, Ibuprofen, celecoxib, gabapentin, fluoxetine, risperidone**
- BP can occur months or sometimes even YEARS after the initiation of these medications
- Must hold possible offending meds x 6 weeks to see if improvement occurs.

MANAGEMENT: STRAIGHT FROM THE DERMATOLOGIST'S MOUTH

- Age appropriate cancer screening should be done if have not already as rarely it can be associated.

Acute Management

- Steroids: High dose steroids Prednisone (0.5mg/kg) or IV Solumedrol in severe cases (1.5mg/kg)
- Continue high dose steroids until no more new blisters then can switch to oral prednisone and taper slowly over several months

MANAGEMENT

If case is more severe (ie large portion of the body is involved, not responding to steroids, immunocompromised patients):

- **IVIG 2 g/kg divided over 2-5 days, then monthly x 3 months**
- **Start on oral Doxycycline 100 mg po od fpr 4 wks**
- **Start on modified cyclosporin 3 mg/kg daily (divided bid) and taper over 3 months (evidence only supports use of this in severe cases where response to steroids alone has not been beneficial)**
- **Apply Betamethasone oint 0.1% all over body daily then cover with abd pads and kling and netting to keep in place.**
- **Lance bullae with sterile needle after cleaning skin but leave roof intact.**

MANAGEMENT

If cases are mild (more likely will be what we see in the office):

- **Potent topical steroid (clobetasol or betamethasone) x weeks to months is FIRST LINE**
- **Stop possible offending agents**
- **May consider oral steroids as well if slightly more severe with a taper over 3 months**
- **Can add oral Doxycycline 100 mg po daily for 4 wks (could be used as first line if contraindications to steroids, but efficacy is not as good)**

MANAGEMENT: OTHER CONSIDERATIONS

- Consider starting something for bone preservation as multiple courses of Prednisone may be needed and most of the bone loss occurs early on (ie. actonel 150 mg per month)
- monitor electrolytes, diabetes (q1-3 months and ensure POC testing at home) and infection
- Biologics may be an option. Dupixent - a biologic used for bullous pemphigoid
- Consult to derm if not improving with the above

BACK TO OUR CASE: A HAPPY ENDING

- After IV steroids, the addition of IVIG and doxycycline, patient finally began to improve, stepped down to oral prednisone
- This allowed him to actually heal and have strength for his Zenker's surgery
- I adopted him as a patient
- I am very happy to say that he has now been started on a biologic, he is off oral steroids,and his skin looks like brand new :)

THANK YOU!